

**333 Brazilian children's perceptions about the routine treatment in cystic fibrosis**

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**Objectives:** The aim of this study was understand the treatment itinerary of Brazilian scholar children with Cystic Fibrosis from their perspective and was supported for the medical anthropology framework.

**Methods:** Data was collected through 15 months following with 7 scholar children, both sex, with CF and their families, with the ethnographic method. Data were analyzed with the hermeneutic framework.

**Conclusion:** CF is part of the life experience of the children and so, it's an important issue of their existence. It has an important role in the decisions of plenty issues like the period of school, the kind of sport that the kid will do and the daily life routine. Most of children did not know clearly what the disease is and nether their treatment and medications names and functions and reported never have been included in his health treatment discussion, what can lead to poor adherence and high dependence to do the daily treatment. Children also have their own explanatory model for the disease and it is different from those of family and CF team, what can lead to truncated communication, understanding and evaluating of the treatment process. Nurses must understand the children perspective about the disease and how their understanding influences their evaluation about the treatment and defines the search for cure. With this we will be able to plan a health care treatment based on the family, children and CF team expectations, with an unified goal that priorities the quality of life instead of the disease control.

**335 Comparing the clinical characteristics of adolescents with CF at transition over the past 10 years**

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**Background:** Most UK CF centres transition adolescents from paediatric to adult care by the age of 18 years. Medical advances and improved transition processes may have changed the characteristics of teenagers moving to adult care now compared to a decade ago.

**Objectives:** To compare the difference in characteristics of teenagers at the time of transition, over a 10 year period with regard to lung function, nutritional status, treatment intensity and age.

**Methods:** Data were collected retrospectively on adolescents at Great Ormond Street Hospital who transitioned to adult care between 2000–2003 (Group 1, n=49) and 2008–2011 (Group 2, n=41). The differences in lung function, nutritional status, microbiological status, treatment intensity and age at transition between the groups were compared.

**Results:** For Group 1, mean (SD) FEV1% predicted and BMI z scores were 68% (22) and -0.46 (1.0) compared to Group 2 of 75% (25) and -0.23 (1.), respectively. Group 1 had a greater proportion of children with chronic *S. aureus* infection ( $p=0.005$ ). Group 2 had a higher proportion receiving Azithromycin ( $p<0.001$ ), nebulised Colomycin and/or Tobramycin ( $p=0.026$ ) and DNase ( $p=0.039$ ). Median age at transition was significantly higher in Group 2 (17.02 compared to 16.6 years,  $p<0.001$ ).

**Conclusion:** Over the past 10 years, there has been a trend towards improvement in FEV1% and BMI z scores, although this did not reach statistical significance. The burden of care for adolescents transitioning today may be greater due to the intensity of the treatment regimen. The older age at transition may be related to the acknowledgement that the transition process requires time and preparation.

**334 What is it like to be an adolescent with cystic fibrosis? Reversing the roles of patient and health-professional**

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**Objectives:** Chronic conditions, like CF, have a significant influence on young patients' quality of life. As these diseases cannot (yet) be cured, it transforms health-care's primary focus from acute treatment to the minimization of risks for complications, and attempts to empower patients to deal with their condition in everyday life. Involving adolescents in their own care proves to be extremely difficult. For example, it is estimated that adolescent patients adhere to only half of their lifestyle and medication regimens in a way that they obtain optimal clinical benefit. In this study, we tried to get more insight into the difficulties of living with a condition like CF.

**Methods:** An explorative, qualitative study. For 4 weeks, 18 health-professionals followed a regimen based on treatment guidelines for adolescent patients with CF, including nebulisation, oral medication, journal keeping, rest and sports activities.

**Conclusion:** The participants' main difficulties were constantly 'feeling different than others' and having to be cautious and illness-aware in every-day situations. Although a motivated and very knowledgeable group of pediatric professionals volunteered to participate they found it surprisingly difficult to adhere to the lifestyle and medication regimen they expected their patients to follow. Recommendations to improve compliance in adolescents with CF are presented.

**336 Neuropsychological functioning in children with cystic fibrosis: First results of a prospective comparative study**

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**Objectives:** The limited number of neuropsychological studies in children with Cystic Fibrosis (CF) focus primarily on intelligence and school performance. Little is known about social cognition and executive functioning (EF), essential for normal development and daily functioning. The aim of this study was to measure social cognition and related functions in children with CF aged 5 to 13 years and to compare these to normative data.

**Methods:** Participants: 20 CF patients from 2 CF centers in the Netherlands. Outcome variables: a standardized neuropsychological battery including tests on social cognition (emotion recognition and theory of mind), EF (planning, working memory and cognitive flexibility) and intelligence; questionnaires on EF completed by parents and teachers; indices of disease severity (Forced Expiratory Volume in 1 second, BMI, exacerbations per year). Analysis: Analysis was performed with nonparametric procedures.

**Results:** Patients' average performance on tests of social cognition, EF and IQ did not significantly differ from normative data. Comparisons to a healthy control group will be presented additionally. Although ratings on EF were in the normal range, teachers indicated significantly more problems than parents on scales of behavioural regulation and metacognition. Disease severity was not significantly related to any of the cognitive variables.

**Conclusion:** Based on this Dutch CF sample, we may conclude that performance on social cognitive, executive and IQ tests is within the normal range and seems unrelated to disease severity. This is in concordance with the sparse publications on other neurocognitive functions in these patients.